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Item 1 of 3
Question Id: 458

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Skin biopsy from the thigh of a child demonstrates vascular lesions with IgA and C3 deposition. The most likely clinical presentation for this patient would be which of the following?

A. Saddle nose and oliguria

B. Skin rash and abdominal pain

C. Weak upper extremity pulses

D. Headache and blurred vision

E. Exertional calf pain and toe ulcers

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Skin biopsy from the thigh of a child demonstrates vascular lesions with IgA and C3 deposition. The most likely clinical presentation for this patient would be which of the following?

A. Saddle nose and oliguria (15%)
B. Skin rash and abdominal pain (74%)
C. Weak upper extremity pulses (2%)
D. Headache and blurred vision (3%)
E. Exertional calf pain and toe ulcers (4%)

Omitted
Correct answer
B

74%
Answered correctly

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02/03/2020
Last Updated

Explanation

Immune complex vasculitis associated with IgA and C3 deposition is typical of Henoch–Schönlein purpura (HSP). A systemic hypersensitivity disease of uncertain etiology, HSP produces leukocytoclastic angiitis in small vessels of the dermis and the gastrointestinal (GI) tract. Clinically, a purpuric rash (100%), colicky abdominal pain (85%), and polyarthralgia (70%) may result. The rash is palpable and usually occurs on the lower extremities at first. Although HSP is usually benign and self-limiting, acute glomerulonephritis is possible complication. The disease is most common in male children, aged 3 to 11.

(Choice A) Nasal mucosal ulcerations (severe ulceration may cause "saddle nose) and glomerulonephritis (producing oliguria) are most characteristic of granulomatosis with polyangiitis (Wegener's). Granulomatosis with polyangiitis is associated with serum cytoplasmic-staining

Immune complex vasculitis associated with IgA and C3 deposition is typical of Henoch–Schönlein purpura (HSP). A systemic hypersensitivity disease of uncertain etiology, HSP produces leukocytoclastic angiitis in small vessels of the dermis and the gastrointestinal (GI) tract. Clinically, a purpuric rash (100%), colicky abdominal pain (85%), and polyarthralgia (70%) may result. The rash is palpable and usually occurs on the lower extremities at first. Although HSP is usually benign and self-limiting, acute glomerulonephritis is possible complication. The disease is most common in male children, aged 3 to 11.

(Choice A) Nasal mucosal ulcerations (severe ulceration may cause "saddle nose) and glomerulonephritis (producing oliguria) are most characteristic of granulomatosis with polyangiitis (Wegener's). Granulomatosis with polyangiitis is associated with serum cytoplasmic-staining antineutrophil cytoplasmic antibodies (C-ANCA), not with IgA.

(Choice C) Weak upper extremity pulses may be found in Takayasu arteritis. This large-vessel vasculitis predominantly involves the aorta and its proximal branches. Histologically, the lesions in Takayasu arteritis are similar to giant cell arteritis.

(Choice D) These symptoms are typical of giant cell arteritis (GCA). GCA is seen in elderly patients. IgA and C3 deposition are absent in GCA lesions.

(Choice E) Distal lower extremity claudication and ulceration would most often be caused by thromboangiitis obliterans or atherosclerosis (ie peripheral vascular disease). Although smoking is a known risk factor for both of these diseases, neither disorders is associated with IgA or C3 deposition.

Educational Objective:
Small vessel leukocytoclastic angiitis associated with IgA and C3 deposition is typical of HSP. HSP is most common in children 3 to 11 years old and is most often related to a recent infection. Most children present with palpable skin lesions, with or without abdominal pain and arthralgias. Although usually self-limiting, patients afflicted with HSP should be observed carefully because glomerulonephritis and even end-stage renal disease are possible complications.

Pathology	Allergy & Immunology	Henoch schonlein purpura
Subject	System	Topic

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Item 2 of 3
Question Id: 11667

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A 21-year-old woman comes to the office due to recurrent episodes of self-limited, colicky abdominal pain. She also had an episode of facial swelling that resolved spontaneously. The patient has no other significant past medical history and takes no medications. Examination is unremarkable. Evaluation shows that her complement protein C1, even when not attached to an antigen-antibody complex, is excessively cleaving C2 and C4. Which of the following is most likely increased in this patient?

A. Antinuclear antibody titer
B. Antistreptolysin O titer
C. Bradykinin
D. Free hemoglobin
E. Renin

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A 21-year-old woman comes to the office due to recurrent episodes of self-limited, colicky abdominal pain. She also had an episode of facial swelling that resolved spontaneously. The patient has no other significant past medical history and takes no medications. Examination is unremarkable. Evaluation shows that her complement protein C1, even when not attached to an antigen-antibody complex, is excessively cleaving C2 and C4. Which of the following is most likely increased in this patient?

A. Antinuclear antibody titer (20%)

B. Antistreptolysin O titer (7%)

✓ C. Bradykinin (61%)

D. Free hemoglobin (7%)

E. Renin (2%)

Omitted
Correct answer
C

61%
Answered correctly

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Time Spent

03/15/2020
Last Updated

Explanation

This patient with recurrent episodes of abdominal pain and an episode of facial swelling likely has angioedema due to **C1 inhibitor (C1INH) deficiency**. C1INH prevents C1-mediated cleavage of C2 and C4, thereby limiting activation of the complement cascade. It also blocks kallikrein-induced conversion of kininogen to **bradykinin**, a potent vasodilator that also causes increased vascular permeability.

Acquired or hereditary **C1INH deficiency** (due to the complete absence of C1INH or the presence of a dysfunctional variant or an anti-C1INH antibody) leads to elevated levels of bradykinin, and patients can develop bradykinin-associated angioedema. Symptoms include facial swelling

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Acquired or hereditary **C1INH deficiency** (due to the complete absence of C1INH or the presence of a dysfunctional variant or an anti-C1INH antibody) leads to elevated levels of bradykinin, and patients can develop bradykinin-associated angioedema. Symptoms include facial swelling (without urticaria), life-threatening laryngeal edema, and gastrointestinal manifestations (eg, nausea/vomiting, colicky pain, diarrhea). Management of acute attacks involves supportive care and the administration of C1INH concentrate or a kallikrein inhibitor.

(Choice A) Increased antinuclear antibody (ANA) titers are seen in a number of autoimmune conditions, including systemic lupus erythematosus, which is associated with hypocomplementemia but would have different clinical manifestations (eg, butterfly rash, arthritis, oral ulcers). ANA production is not a result of excessive C1 activity.

(Choice B) Poststreptococcal glomerulonephritis is associated with increased antistreptolysin O titers, complement activation, and low levels of C3. The typical presentation is hematuria or nephritic syndrome following respiratory infection.

(Choice D) Complement-mediated intravascular hemolysis can result from autoantibodies (autoimmune hemolytic anemia) or from direct complement activation (paroxysmal nocturnal hemoglobinuria); however, neither condition results from excessive C1 activity.

(Choice E) Angiotensin-converting enzyme (ACE) inhibitors, which are associated with idiopathic angioedema, function by blocking ACE and result in increased renin levels. ACE also degrades bradykinin.

Educational objective:
C1 inhibitor (C1INH) deficiency causes increased cleavage of C2 and C4 and results in inappropriate activation of the complement cascade. C1INH also blocks kallikrein-induced conversion of kininogen to bradykinin, a potent vasodilator associated with angioedema.

References

- C1 inhibitor deficiency: consensus document.
- Hereditary angio-oedema.
- Pathophysiology of hereditary angioedema

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Item 3 of 3
Question Id: 459

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A 43-year-old man is hospitalized with recent-onset oliguria and a high serum creatinine level. He has been seen in clinic several times for an intranasal ulcer that has failed to heal. This patient's condition is most likely associated with antibodies against which of the following?

A. Glomerular basal membrane
B. Smooth muscle cells
C. Neutrophils
D. Erythrocytes
E. Platelets
F. Mitochondria

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A 43-year-old man is hospitalized with recent-onset oliguria and a high serum creatinine level. He has been seen in clinic several times for an intranasal ulcer that has failed to heal. This patient's condition is most likely associated with antibodies against which of the following?

- A. Glomerular basal membrane (35%)
- B. Smooth muscle cells (6%)
- ✓ C. Neutrophils (49%)
- D. Erythrocytes (0%)
- E. Platelets (5%)
- F. Mitochondria (2%)

Omitted
Correct answer
C

49%
Answered correctly

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03/21/2020
Last Updated

Explanation

Nasal mucosal ulcerations and glomerulonephritis are most characteristic of granulomatosis with polyangiitis (Wegener). Cytoplasmic-staining antineutrophil cytoplasmic antibodies (c-ANCAs) are virtually pathognomonic for granulomatosis with polyangiitis, with a better than 90% specificity and sensitivity. Proteinase 3-ANCA (also known as c-ANCA) may also be useful as a quantitative measure of disease activity.

(Choice A) Goodpasture syndrome is sometimes called anti-glomerular basement membrane (GBM) antibody disease. Presenting clinical manifestations of Goodpasture syndrome are hemoptysis, radiographic focal pulmonary consolidations, and a glomerulonephritis that may rapidly progress to renal failure. Upper respiratory tract ulceration is not characteristic.

Explanation

Nasal mucosal ulcerations and glomerulonephritis are most characteristic of granulomatosis with polyangiitis (Wegener). Cytoplasmic-staining antineutrophil cytoplasmic antibodies (c-ANCAs) are virtually pathognomonic for granulomatosis with polyangiitis, with a better than 90% specificity and sensitivity. Proteinase 3-ANCA (also known as c-ANCA) may also be useful as a quantitative measure of disease activity.

(Choice A) Goodpasture syndrome is sometimes called anti-glomerular basement membrane (GBM) antibody disease. Presenting clinical manifestations of Goodpasture syndrome are hemoptysis, radiographic focal pulmonary consolidations, and a glomerulonephritis that may rapidly progress to renal failure. Upper respiratory tract ulceration is *not* characteristic.

(Choice B) Smooth muscle cell antibodies are commonly seen in patients with autoimmune hepatitis.

(Choice D) Antierythrocyte antibodies can produce hemolysis of an intravascular and/or extravascular type. Hemoglobinemia from intravascular hemolysis can end in acute renal failure, although this is predominantly seen after transfusion reactions only when other conditions, such as hypovolemia and/or acidemia, coexist. Hemolytic anemia would not cause isolated ulceration of mucosa.

(Choice E) Platelet antibodies can often cause thrombocytopenia by binding to platelets. Neither these autoantibodies, nor thrombocytopenia is known to cause acute renal failure.

(Choice F) Antimitochondrial antibodies are seen in primary biliary cholangitis (cirrhosis).

Educational objective:
Necrotizing vasculitis of the upper and lower respiratory tract (causing nasal ulcerations, sinusitis, hemoptysis) and rapidly progressive glomerulonephritis—producing a variable degree of renal failure—is characteristic of granulomatosis with polyangiitis (Wegener). This disease is associated with C-ANCAs, which may target neutrophil proteinase 3.

Pathology	Allergy & Immunology	Granulomatosis with polyangiitis
Subject	System	Topic

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